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# A rare case of Heyde's syndrome with non-classic presentation

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## Abstract

Heyde's syndrome is characterized by gastrointestinal bleeding due to colonic angiodysplasia associated with aortic stenosis. This association does not explain the pathophysiology, but an occasional coincidence in the elderly. We present a non-classic case associated with left ventricular outflow obstruction and steal phenomenon.

## Keywords

Heyde's syndrome; aortic stenosis; LVOT obstruction; angiodysplasia

## Case Report

A 71-year-old woman, smoker with a history of Hypertension, Sickle cell anemia, Carpal Tunnel syndrome, Restrictive lung disease, PUD, Diverticulosis, Mitral Regurgitation, and Left Ventricular Outflow Tract Obstruction was admitted to the hospital with a 1-day history of weakness and black tarry stools. Of note, the patient has multiple admissions with bleeding per rectum.

On physical exam, she was tachycardic. She was noted to have conjunctival pallor, diminished second heart sound, left carotid bruit. Her hemoglobin was 6.9 g/dl. Platelets, international normalized ratio, prothrombin time, and partial thromboplastin time were within normal limits. Chest X-ray was normal. CT chest angiography reveals no aortic dissection.

After 2 units of blood were transfused, and after stabilization, EGD and colonoscopy were performed and only angiodysplasia found in the descending colon and was suspected to be the source of the bleeding. Echocardiography later showed severe hypertrophic cardiomyopathy with severe asymmetric septal hypertrophy and severe narrowing of the left ventricular outflow tract. On CT angiography the patient severe focal stenosis of subclavian artery correlating with subclavian steal syndrome. With the recurrent episodes of obscure gastrointestinal bleeds and the examination findings suggestive of LVOT obstruction, Heyde's syndrome was suspected associated with subclavian steal syndrome in our patient.

## Discussion

The relationship between the aortic outflow obstruction and lower gastrointestinal bleeding is not well studies and is not well known till now, and all the explanation is built on assumptions. In patients over 60 years of age, both calcific AS and AD have to be considered as the result of degenerative vascular changes [1,2].

Boley et al. believe that the prevalence of angiodysplasia at the ascending and transverse colon is due to the higher intraluminal pressure resulting from the larger diameter of this part of the colon. Consequently, the veins draining the mucosa and perforating the muscular layer of the colon wall become obstructed, tortuous and finally form arterio-venous malformations [1].

Weaver et al. suggested that a connective tissue defect, might cause both the changes of the aortic valve (calcific degeneration) and dilatation of the blood vessels of the colon, in other words they both have the same cause [4]. Also, due to low cardiac output, AS might cause ischemia of the mucosal layer of the colon and thus local endothelial necrosis in the angiodysplastic areas and bleeding [1,3,4], However, this has not been confirmed yet [4].

Still the question why is the bleeding, one of the most accepted theories is the shearing mechanism and its effect on the Von- Willebrand factors. The theory stated that when the bloodstream passes through narrowed Aortic valve or LVOT obstruction, it shears and leads to consumption of WVF, and leads to more bleeding in the area of the Angiodysplasia in the colon which is the weak area. That might explain why the bleeding ceases after aortic valve replacement or ablation to the hypertrophied LV wall with alcohol injection [5].

Von Willebrand factor (vWF) synthesized by endothelium is stored in ultra-large multimers and mediates platelet adhesion at sites of vascular damage [6,7]. It circulates as large multimers, each one exceeds the size of a platelet [8,9]. The high velocity of blood flow within telangiectasia requires the largest multimers of vWF to maintain hemostasis [10].

A product called acquired vWS-2A in aortic stenosis or LVOT obstruction arises from degradation of vWF multimers by the shear mechanism across the narrowed passages whether aortic stenosis, severe hypertrophic hypertrophy of stenosis on one of the big arteries. An aortic pressure gradient of 50 mmHg may cause coagulopathy [11]. vWF multimers unfold from a coiled structure to an elongated filament and are then cleaved [12-15]. Degradation of the heaviest multimers of vWF, which are responsible for platelet-mediated hemostasis, is the culprit for coagulopathy in Heyde's syndrome, which again is the most accepted theory [13].

In a study of 50 consecutive patients with aortic stenosis, aortic valve replacement (AVR) treat 84%, 21% of patients with severe aortic stenosis suffered from cutaneous or mucosal bleeding, and 67–92% had other hematological abnormalities which correlated with the severity [13].

VWF was measured at 1 day, 7 days, 6 months after the procure of AVR. vWF level was corrected on the 1st day after intervention but tends to happen again at 6 months [13].

Yoshida et al. showed electrophoretic deficits of large multimers of vWF in patients with aortic stenosis which resolved postoperatively, but no differences in pre- and postoperative vWF were found in patients with severe mitral regurgitation [16].

Our patient was discharged home after endoscopic coagulation to follow up with the cardiologist for possible ablation of the hypertrophic LV wall, and follow up with the primary care for bleeding studies and possible recurrent attacks of bleeding till definite treatment.

## Conclusion

Heyde's Syndrome is a rare occurrence that is shown to present with gastrointestinal bleeding due to colonic angiodysplasia associated with aortic stenosis. Interest appears due to the pathophysiologys' inability to substantiate the occurrence. This patient had a rare case since she presented with left ventricular outflow tract obstruction and subclavian stenosis in addition to gastrointestinal bleeding. Treatment remains conservative in mild cases of gastrointestinal bleeding and in severe or recurrent cases may require aortic valve replacement or ablation to the LV hypertrophic wall, with semi-annual follow ups. Due to the syndromes elusive source, it is imperative that researchers continue to investigate until a diagnosis can be confirmed through an associated direct cause.

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